

A case of pediatric paraparesis secondary to an idiopathic acute transverse myelitis

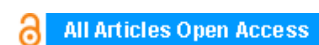
Joana Teixeira, Susana Carvalho, Sofia Martins, Teresa Pontes, Alvaro Machado, Henedina Antunes

ABSTRACT

Introduction: Acute transverse myelitis (ATM) refers to a frequently idiopathic, segmental spinal cord inflammation. It is a rare condition, in particular in children, and not previously reported in a family retinitis pigmentosa (RP) clinical setting.

Case Report: An 11-year-old previously healthy girl, with a family history of RP, presented with a subacute flaccid paraparesis, with bilateral, up to the fourth dorsal level, mixed sensory hypoesthesia and autonomic dysfunction. Brain and spinal cord magnetic resonance imaging (MRI) showed an extensive, T2-hyperintense, non-contrast enhancing lesion from the second to fifth dorsal levels. Cerebrospinal fluid (CSF) and lab studies were normal, as the ophthalmologic observation. Treated with high-dose corticosteroids and intensive physical therapy, a significant recovery could be seen.

Conclusion: Early pharmacological and physical treatment is fundamental and may indeed change the prognosis of this disease ATM. The family history of RP, although probably incidental, brings nevertheless the issue of a possible etiological contribution, or pathologic common pathways.



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CASE REPORT

OPEN ACCESS

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Introduction: Acute transverse myelitis (ATM) refers to a frequently idiopathic, segmental spinal cord inflammation. It is a rare condition, in particular in children, and not previously reported in a family retinitis pigmentosa (RP) clinical setting. **Case Report:** An 11-year-old previously healthy girl, with a family history of RP, presented with a subacute flaccid paraparesis, with bilateral, up to the fourth dorsal level, mixed sensory hypoesthesia and autonomic dysfunction. Brain and spinal cord magnetic resonance imaging (MRI) showed an extensive, T2-hyperintense, non-contrast enhancing lesion from the second to fifth dorsal levels. Cerebrospinal fluid (CSF) and lab studies were normal, as the ophthalmologic observation. Treated with high-dose corticosteroids and intensive physical therapy, a significant recovery could be seen. **Conclusion:**

Early pharmacological and physical treatment is fundamental and may indeed change the prognosis of this disease ATM. The family history of RP, although probably incidental, brings nevertheless the issue of a possible etiological contribution, or pathologic common pathways.

Keywords: Transverse myelitis, Retinitis pigmentosa, Paraparesis, Neurogenic urinary bladder

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INTRODUCTION

Acute transverse myelitis (ATM) refers to a multiple-level segmental spinal cord injury, caused by an acute inflammatory process.

It is very rare, with an estimated incidence of 1–5 cases per million per year [1]. Of these, only 1/5 occur in children, mainly before the age of two (a bimodal incidence can be seen, with a low number of cases between two and five years) [1, 2].

Although commonly idiopathic, an autoimmune disturbance is frequently suspected, and a polyphasic demyelinating disorder can only be disregarded after a reasonable follow-up period.

Clinically, it is characterized by acute to subacute onset of a variable signs of motor, sensory and autonomic dysfunction, which can be localized to a certain level (commonly a series of adjacent levels) of the spinal cord.

It can have major consequences, with residual sensitive, autonomic and motor dysfunction in up to 20% of cases [3].

Retinitis pigmentosa (RP) refers to a heterogeneous group of inherited ocular diseases resulting in a progressive retinal degeneration. It affects 1 in 3,000–5,000 people and occurs in isolation or in a syndromal manner [4, 5].

CASE REPORT

An 11-year-old previously healthy gymnastics practitioner girl, with anisometropia and family history of RP (mother and maternal aunt), was seen for a 4-day-evolving lower-limb loss of strength and sensitivity, combined with dorsal pain and sphincter dysfunction.

There was no fever or recent relevant traumatic injury.

Neurological examination revealed a left-predominant flaccid paraparesis with normal myotactic and superficial reflexes, a mixed sensory disturbance with algic hypoesthesia up to the fourth dorsal level, and proprioceptive distal loss.

Magnetic resonance imaging (MRI) of the medulla showed slight dorsal high intensity signal in all T2-weighted sequences. Laboratory examination, including all virologies, relevant serologies and immunity screening, were found to be normal. Cerebrospinal fluid (CSF) was also completely normal (including absent oligoclonal bands), as it was the computed tomography (CT) angiography of chest and neck vessels.

Assuming the most likely diagnosis of ATM, she was admitted and started methylprednisolone bolus (30 mg/kg/day).

MRI was repeated at day-12 showing that the signal change extended from the first to fifth dorsal level, was bright in all T2-weighted sequences, did not uptake administered gadolinium, and was more clearly localized in the lateral and anterior columns (Figure 1).

At ophthalmologic evaluation there was no evidence of changes in visual acuity or ocular fundus.

Clinical improvement started at day-5 of methylprednisolone. There was increase muscle strength sufficient for autonomous, although limited, deambulation. It was decided to keep corticosteroid therapy (oral prednisolone 1 mg/kg/day). She also started physiatrist treatment with further muscle strength improvement and gait control. Bladder catheterization was needed because of high post-voiding residual volume.

She was discharged at day-23, with residual paraparesis (Medical Research Council Scale grade 4+), maintaining prednisolone, physiatrist treatment and intermittent bladder catheterization.

Five months later MRI was repeated, showing a reduced dorsal medulla thickness (Figure 2).

Currently, six months after, she maintains paraparesis, being capable of walking for short distances, but needing help for longer distances, personal hygiene and clothing. Neurogenic bladder persists, with secondary nocturnal enuresis, but with spontaneous daytime voiding.



Figure 1: MRI (sagittal section), T2-weighted sequences, showing a first to second dorsal level hyperintense signal, extending to the fourth to fifth dorsal level, more clearly localized in the lateral and anterior columns, with no uptake of administered gadolinium.

DISCUSSION

In ATM, symptoms are grouped on physiological ground as motor, sensory or autonomic. They greatly vary because of the topographical variability (the level, extension and localization) of the disease and, whenever secondary, of the pathology of the underlying cause. As so, clinical onset can be rapidly progressive or more slowly-evolving over a few weeks, and symptoms can affect all limbs, only the lower ones, with or without symmetrical impairment, and predominantly affect one of the three above-mentioned motor, sensory and autonomic systems [2]. MRI is the fundamental study to carry in an emergency setting so to exclude compressive lesions. In ATM,



Figure 2: MRI (sagittal section), T2-weighted sequences, showing a hyperintense signal of anterior dominance in the middle dorsal region, associated with a reduced dorsal medulla thickness.

lesions are found mainly in the white matter surrounding the central medullar channel, usually involving several adjacent medullar segments, and are more easily seen in T2-weighted sequences, where the edema appears bright. This usually precedes the latter medullar atrophy, the sole imagiological evidence of a past ATM [6–8].

In the majority cases, CSF has increased protein content as well as mild lymphocytosis. However, as spinal cord inflammation may not be evident at the beginning, some authors suggest that lumbar puncture should be repeated between second day and seventh day of the disease [6].

Oligoclonal bands should always be sought, because if they are found in the CSF and not in the blood, they raise the risk for multiple sclerosis [9]. An ophthalmologic evaluation is also recommended for all patients, as an additional finding of optic neuritis has a major implication on the diagnosis (multiple sclerosis or optic neuromyelitis) [6, 9].

Treatment is not consensual. The first line therapy is methylprednisolone for 5–7 days, followed by oral prednisolone (1 mg/kg/day) for 3–4 weeks. Non-pharmacological treatment includes intermittent bladder catheterization and physiotherapy [2, 9].

Etiological considerations in ATM should include viral/bacterial infections, autoimmune and connective tissue diseases, demyelinating diseases (multiple sclerosis, neuromyelitis optica), intra or extra-axial tumors and vascular diseases. Regarding the last etiological group idiopathic spontaneous dorsal spinal cord infarction is another possible unusual etiology of acute paraparesis in children. When it is possible to exclude all of these causes, ATM is then classified as idiopathic [1, 2, 6].

No etiologic cause of the ATM was found. Given the family history of (RP), a possible association was raised, although both the absence of prior descriptions and of RP signs in our patient, strongly reduce this possibility. There are few descriptions of RP associated with central nervous system infections. It was related to Creutzfeldt-Jakob disease and with congenital toxoplasmosis [4, 10]. There is also been described a close association between Human T-lymphotropic virus type I (HTLV-I) infection associated myelopathy and RP although the pathogenesis remains to be defined [5]. On this ground, we raise the possibility of an etiological contribution, or pathologic common pathways between ATM and RP.

CONCLUSION

Early pharmacological and physical treatment is fundamental and may indeed change the prognosis of this disease ATM. The family history of RP, although probably incidental, brings nevertheless the issue of a possible etiological contribution, or pathologic common pathways.

Author Contributions

Joana Teixeira – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Susana Carvalho – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Sofia Martins – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Teresa Pontes – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Álvaro Machado – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Henedina Antunes – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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